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Factors influencing the 6-min walking test (6MWT) in Cystic Fibrosis (CF)

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Background: The 6-minute walking test is a popular, practical, simple clinical exercise test that provides a global assessment of functional capacity.
Purpose: To identify factors influencing the 6MWT performance in stable adolescent and adult CF patients.
Methods: Twenty- six adolescent and adult CF patients having stable clinical condition (13 M, age: 20, 9 ± 5,7) and mild to moderate lung disease (FVC 81±25 %pr, FEV_1 70±26 %pr and FEF_{25-75} 49±35 %pr) were enrolled. Six- minute walking distance (D) in meters was measured in all patients. All of them underwent lung HR-CT, pulmonary function tests (lung volumes, flow rates and diffusing capacity) and arterial blood gases evaluation. Maximal Inspiratory (P_{Imax}) and Expiratory (P_{Emax}) muscle Pressures were also recorded. Unattended sleep studies with the Embletta portable system were performed the same night. Body mass index (BMI) was calculated and serum values of albumin, ferritin and transferin were measured.
Results: Statistically significant correlations between D and min SpO2 in overnight oximetry ($p<0.002$), DLCO % ($p<0.02$), heart rate in exercise ($p<0.03$) were found. A negative correlation was found with total HR-CT score and RV/TLC ($p<0.04$ and 0.02 respectively). No correlations between D and serum factors of nutritional status were found.
Conclusions: Six minute walking distance is a good indicator of the functional status in stable CF patients, as proved by common functional and radiographic tests.

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Cardiac function in adolescent and adult cystic fibrosis (CF) patients

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Background: Subclinical pulmonary hypertension is considered to be associated with an increased mortality in CF. It is thought to be related to progressive destruction of the lung parenchyma and vasculature and to pulmonary vasoconstriction secondary to hypoxemia.
Purpose: To determine the prevalence of subclinical pulmonary hypertension and cardiac dysfunction in stable adolescent and adult CF patients and to examine the relationship between cardiovascular abnormalities, lung function and hypoxemia.
Methods: Twenty-two adolescent and adult CF patients in stable clinical condition (11 M, mean age 20,5 ± 5,4) and mild to moderate lung disease (FVC 82 ± 26%, FEV_1 67 ± 27% and FEF_{25-75} 47 ± 35%pr) were consecutively enrolled. All of them underwent PFTs (lung volumes, flow rates and diffusing capacity), arterial blood gases evaluation and 6 min walking test (6MWT). Maximal Inspiratory (P_{Imax}) and expiratory (P_{Emax}) muscle pressures were recorded. M-mode, 2-D and Doppler echocardiography as well as overnight unattended sleep study with the Embletta portable system were performed the same day.
Results: Nobody of the examined subjects had developed pulmonary hypertension. Correlations were found between: 1.RightVentricularEnd-Diastolic and Left Atrium diameter ($p<0.001$) as well as Pittsburg Insomnia Scale ($p<0.009$) 2. Inter-ventricular Septum Thickness with systolic Blood Pressure ($p<0.02$) 3.Left Ventricular End-Diastolic Diameter with systolic Blood Pressure ($p<0.02$) and Heart Rate at rest ($p<0.016$).
Conclusions: In adolescent and adult stable CF patients having mild to moderate lung disease left and right ventricular function is not affected.

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Inspiratory capacity as a predictor of exercise tolerance in adult patients with cystic fibrosis

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INTRODUCTION: The aim of this study was to examine the role of inspiratory capacity as a predictor of oxygen kinetics during maximal exercise and early recovery in adult patients with cystic fibrosis (CF).
PATIENTS AND METHODS: Eighteen patients with CF and eleven healthy subjects underwent resting pulmonary function testing, including inspiratory capacity (IC) and symptom-limited treadmill cardiopulmonary exercise testing (CPET). For CF group percentage of predicted FEV_1 was 76.8 ± 32.8, percentage of predicted FVC was 95.0 ± 29.6, FEV_1/ FVC was 68.9 ± 16.1 and percentage of predicted IC was 82.4 ± 35.9. Mean peak oxygen uptake (VO_2 peak) was 29.1 ± 7,0 ml/kg/min and VO2/t slope was 0,59 ± 0,25.
RESULTS: In a multivariate stepwise regression analysis, using VO_2 peak as the dependent variable and the pulmonary function test measurements as independent variables, the only significant predictor selected was IC ($r=0.61$, $p<0.007$), respectively using VO_2/t slope as a dependent variable, the only significant predictor selected was IC ($r=0,86$, $p<0.0001$). In a final stepwise regression analysis including all the independent variables of resting pulmonary function tests, the only predictor selected was IC ($r^2= 0.37$ and $r^2 = 0.74$ respectively for VO_2 peak and VO2/t slope).
CONCLUSION: In patients with cystic fibrosis, IC inversely related to VO_2 peak and VO2/t slope and is a strong independent predictor of exercise capacity.

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The effects of a multimodality treatment on respiratory muscles and clinical indices in adult patients with cystic fibrosis

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INTRODUCTION: Cystic fibrosis (CF) is characterized by recurrent infections. The strength of respiratory muscles is related to the effectiveness of cough and mucociliary clearance.
This study was devised to assess the effect of a long-term out-patient combination physiotherapy along with respiratory muscle training with the spirotiger for a year, on Schwachman Score (SS) and the strength of respiratory muscles.
PATIENTS AND METHODS: In 15 CF patients (7male/8female) aged 25±9, FEV1 55±35 % of predicted and SS 65±12, maximal inspiratory (Pimax) and maximal expiratory (Pemax) pressure were measured at the onset and after 4-6 months of the multimodality treatment: physiotherapy (autogenic drainage and forced expiration technique) and respiratory muscle training with the spirotiger for a year. For the measurement of Pimax, each patient was instructed to exhale to RV and then to make a maximal inspiratory effort through a closed mouthpiece. For the measurement of Pemax, each patient was instructed to inhale to TLC and then to make a maximal expiratory effort through a closed mouthpiece. The highest peak pressure of the three attempts that differed less than 5% was chosen.
RESULTS: After treatment there was improvement in Pimax (85,2±34,6 vs 71,46±36,7 cmH_2O, $p < 0,042$) and in Pemax (100,3±59,3 vs 75±37,1 cmH_2O, $p < 0,046$) compared with the initial measurements. SS also improved (74±15 vs 67±14, $p < 0,003$) whereas there was no improvement in FEV_1 (65±39 vs 58±35 % of predicted, $p < 0,18$).
CONCLUSION: These results indicate that the aforementioned multimodality treatment (physiotherapy and respiratory muscle training with the spirotiger) improves the strength of respiratory muscles in patients with cystic fibrosis.